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Waterhouse-Friderickson Syndrome

Recovery of a Patient Treated with Cortisone

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THE clinical entity of severe meningococcemia with purpura, cyanosis and shock, was until recent years always fatal. Since the advent of penicillin, sulfonamides and adrenocortical extract, a few cases in which the patient recovered have been reported.

The cause of death in this disease is damage to the adrenal glands, either from gross hemorrhage into the organs or degeneration of them as a result of the overwhelming sepsis.

Since the discovery of cortisone, reports of several cases in which the drug was successfully used in treatment have been published.^{1, 2}

The following case is reported because it was so severe that the patient probably would have died save for cortisone therapy.

REPORT OF A CASE

A three-year-old white female was admitted to St. Mary's Long Beach Hospital on June 8, 1951, in critical condition. Twelve hours prior to admission, high fever and headache had developed. The patient had spent a restless night and had vomited once. Six hours after the onset of fever, a rash appeared over the body and extremities. At first erythematous and discrete, it gradually became confluent and purpuric. The high fever continued and the child gradually became comatose.

When examined upon admission, the patient was stuporous but could be aroused by stimulation. Cyanosis of the lips and nail beds was noted. The skin was cold and clammy. Petechial and ecchymotic eruptions, the largest 3x4 cm. in diameter, covered the body and extremities. The pulse was barely perceptible and the rate could not be determined. The rectal temperature was 105° F. The patient appeared to be moribund. There was no local or general glandular enlarge-

ment. Both ear-drums were intact and normal. A purulent postnasal drip was noted, and the capillary vessels in the pharynx were distended. The pupils were round, regular and equal and reacted to light. No abnormality was noted in percussion and auscultation of the lungs. Percussed, the heart seemed to be normal in size. In stethoscopic examination the sounds were very faint and the pulsation rapid. The rate could not be counted. A reading of the blood pressure was unobtainable. The abdomen was soft and non-tender. The tip of the spleen was palpable. The external genitalia were normal in appearance. The deep reflexes were hypoactive. Superficial reflexes were present. Kernig's sign and Brudzinski's sign were elicited.

An x-ray film of the chest was essentially normal. Except for a pulse rate of 166 to 188 per minute, no abnormality was noted in an electrocardiogram. Erythrocytes numbered 3,640,000 per cu. mm. and the hemoglobin content was 10 gm. per 100 cc. Leukocytes numbered 16,400 with a normal cell differential. The platelet count was 185,640. Many hyaline and fine granular casts were noted in examination of the urine. In the spinal fluid there were 10 lymphocytes per cu. mm. The sugar content was 50.1 mg. per 100 cc. and the protein content 35 mg. No organisms were observed in a smear of the fluid and none grew on a culture.

Gram-negative diplococci grew on a culture of blood taken at the time of admission.

The patient was placed in an oxygen tent and given 25 gm. of cortisone intramuscularly every four hours. Four hundred thousand units of penicillin in aqueous solution and 1 gm. of streptomycin were given intramuscularly immediately, and an infusion of 5 per cent glucose solution was started. Sulfadiazine, 0.1 gm. per pound of body weight per day, was given by mouth several hours later when the patient was able to take fluids by mouth. Four hours after admission the patient's color improved, the blood pressure was 70 mm. of mercury systolic and 40 mm. diastolic, and the pulse rate was 130 per minute. During the next 12 hours there was gradual improvement in the patient's condition and the cyanosis cleared. Small amounts of fluids were taken by mouth. The blood pressure rose to 80 mm. of mercury systolic and 50 mm. diastolic. The rash became more erythematous except for the large ecchymotic areas. For the next three days the patient gradually improved further. The temperature returned to normal and the oxygen was discontinued. On the fifth hospital day the average number of circulating eosinophils was 88.8 per cu. mm. and cortisone was discontinued. The rash gradually disappeared and the patient was discharged on the tenth hospital day.

SUMMARY

A three-year-old child with a severe case of Waterhouse-Friderickson syndrome recovered following treatment with cortisone.

It is believed that cortisone is a valuable aid in treating patients with the disease and in tiding them over the critical phase of the disease.

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